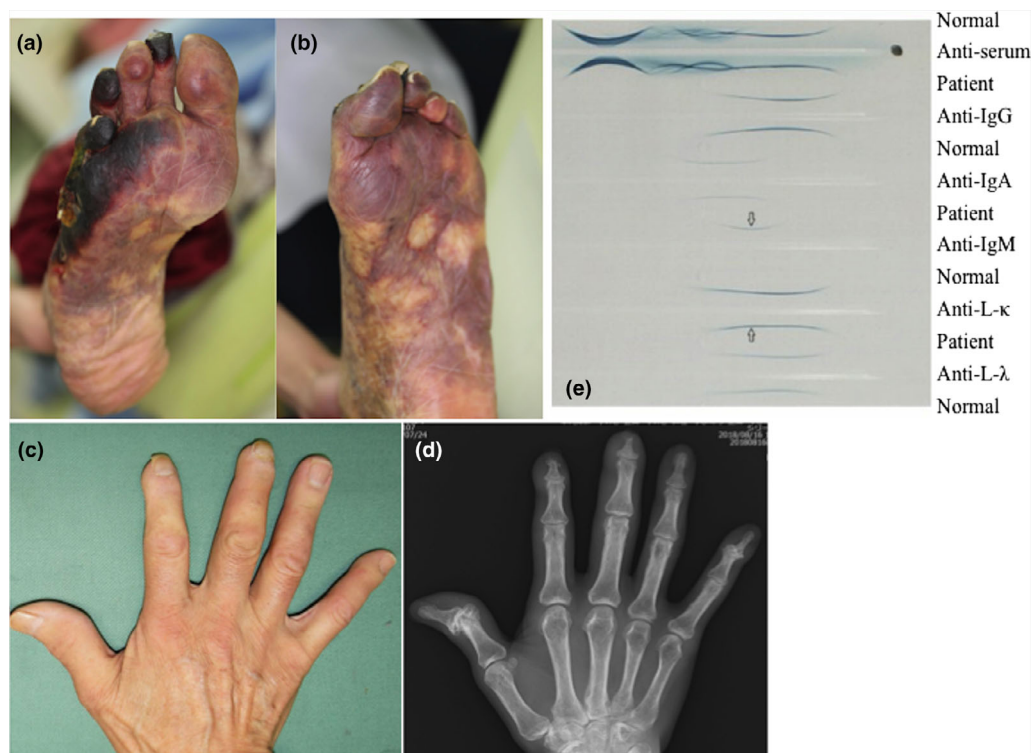


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### Clinical Images: Digital necrosis due to cryoglobulinemic vasculitis secondary to scleroderma




**Figure 1.** The findings of cryoglobulinemic vasculitis associated with scleroderma

A 71-year-old woman whose anticentromere antibody-positive limited cutaneous systemic sclerosis had been treated with mycophenolate and antiplatelet presented with a 4-week history of malaise and progressive discoloration and pain in the digits this winter. Physical examination revealed blue-black discoloration of the distal left first through fifth toes, dusky discoloration of several right distal toes, and purpuric lesions on soles (Figure 1A and 1B). There was sclerodactyly and telangiectasia limited to fingers with osteolysis of distal phalanx (Figure 1C and 1D). Bilateral posterior tibial and dorsalis pedis artery pulses were palpable. There had been no response to anticoagulant and vasodilator therapy for 2 weeks, and then the necrosis got worse. Laboratory tests showed normal renal and hepatic function. C-reactive protein level was elevated at 7.45 mg/dl (normal value <0.3). Digital necrosis developed in cold days (ie, winter) has a broad differential diagnosis, including vasculitis, infection, arterial embolism, and thrombophilia. Results of testing for antineutrophil cytoplasmic antibodies, antinuclear antibody, antiphospholipid antibodies, hepatitis virus, and cold agglutination were negative, but cryoglobulinemia was revealed. Immunoelectrophoresis showed immunoglobulin M (IgM) κ-type M-protein (Figure 1E, arrow). Additional findings showed hypocomplementemia (CH50, <10.0 U/ml; C4, 3.0 mg/dl). Bone marrow biopsy showed no findings of multiple myeloma. There were no growth on blood cultures, and transesophageal echocardiography showed no valvular vegetations. The patient was clinically diagnosed with cryoglobulinemic vasculitis (CV) secondary to scleroderma. Treatment with prednisolone was initiated, and rituximab was later added for early decrease of prednisolone. Biopsy of the digital lesions after initiation of glucocorticoid therapy demonstrated inflammation and medial thickening in a small-sized artery with C4 and IgM deposition, confirming diagnosis of necrotizing vasculitis. Following treatment, symptoms improved, and C-reactive protein level normalized. Necrotic areas of the toes were amputated, but all other digits recovered completely. CV secondary to scleroderma is rare; however, digital necrosis that develops in winter could be one manifestation of scleroderma with cryoglobulinemia (1). This case should remind readers to consider CV as a cause of acute digital necrosis in patients with scleroderma.

Author disclosures are available at <https://onlinelibrary.wiley.com/action/downloadSupplement?doi=10.1002%2Facr2.11395&file=acr211395-sup-0001-Disclosureform.zip>.

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